Towards the optimal Fontan operation: a single-institution experience

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Abstract

Many groups, including ours, have played a more or less significant role in the development and evolution of the Fontan operation that has taken place over the past four decades. This is a brief review of the contributions developed by our institution. Contributions to clinical and technical areas include: development of the ventricular exclusion concept; promoting the staged approach to the Fontan operation by bidirectional cavopulmonary anastomosis with or without a modified Damus—Kaye—Stansel anastomosis; endorsement of the total extracardiac cavopulmonary connection; a rational approach to the management of systemic venous aberrances in single-ventricle patients; and criteria for transition to heart transplantation. Contributions to computational fluid dynamics include: investigation of the best spatial arrangement for the total extracardiac cavopulmonary anastomosis and identification of the so-called 'beneficial vortex'; studies on the effect of unbalanced pulmonary resistances on the hydrodynamic performance of the total extracardiac cavopulmonary connection; and a rationale for the management of systemic venous anomalies in single-ventricle patients.

Keywords bidirectional cavopulmonary anastomosis; Fontan operation; total cavopulmonary connection

Since its introduction for the treatment of tricuspid atresia in 1968, the Fontan operation has evolved into the universal surgical solution for the entire spectrum of functionally univentricular conditions, as well as for some very complex biventricular conditions.[1]

The historical development of the Fontan operation can be divided into three eras (Figure 1). The first era, between 1950 and 1975, focused on theorisation of the 'dispensable right ventricle' concept[2] and culminated in clinical application of the *Glenn shunt* (1958)[3] and the *atriopulmonary anastomosis* (*Fontan operation*; 1968—1971).[1] The second era (1975—1990) concentrated on the expansion of the original atriopulmonary connection model and its upgrade into the hydraulically more efficient *total cavopulmonary connection* model.[4] Finally, the third era (1990 to the present day) has involved optimisation of the total cavopulmonary connection and a search for its best spatial arrangement.

Over the last 25-year period, our group has actively participated in the quest for the ideal technical assemblage and clinical utilisation of the Fontan circulation with a number of both clinical and computational fluid dynamic studies. The following is a brief review of the contributions developed at Bambino Gesù Children's Hospital in Rome, Italy, which started around the middle of the second 'Fontan era'.

Clinical and technical contributions

Ventricular exclusion

Shortly after the introduction of the Fontan operation for tricuspid atresia, it became increasingly evident that the use of the atriopulmonary connection could be extended to the surgical treatment of a variety of otherwise anatomically uncorrectable univentricular[5,6] and occasionally biventricular[7] lesions (the modified Fontan operation). This was made possible by the addition of the so-called *ventricular exclusion* procedure, i.e. the combined obliteration of the atrial communication and of the right-sided atrioventricular valve. Originally, we anchored the occluding patch directly to the leaflets of the right atrioventricular valve with interrupted mattress sutures placed 1 mm away from the valve annulus. However, late dehiscence of the patch occurred in 3 of 5 patients treated by this technique. Therefore, it was decided to suture the ventricular exclusion patch onto the structurally more resilient atrial wall, about 5 mm away from the annulus of the right atrioventricular valve. This manoeuvre allowed the atrioventricular node to be avoided and exiled the coronary sinus onto the ventricular side of the patch but above the right atrioventricular valve, without haemodynamic consequences.[8]

Staging towards a Fontan operation

The results of initial surgical series of modified Fontan operations carried an unexpected rate of attrition at repair despite adherence to the classical '10 commandments' of Fontan and Baudet.[5,9] It was then realised that the circulatory pattern of all univentricular hearts, whether in their natural history or after a neonatal palliation, is one of recirculating parallel pulmonary and systemic flows. The ensuing chronic volume overload may induce progressive ventricular dilatation with increasing biomechanical wall stress and potentially irreversible changes in myocytic structure. The sudden application of a Fontan operation, which abruptly removes the excessive load, may determine an unfavourable geometric adjustment of the systemic ventricle that ultimately results in relative ventricular hypertrophy and secondary diastolic dysfunction.[10]

Somewhat empirically, borrowing from previous works of Haller et al.,[11] Azzolina et al.,[12] Hopkins et al.[13] and Kawashima et al.,[14] in 1985 we promoted the use of *bidirectional cavopulmonary anastomosis* (BCPA) as a staged or definitive palliation in candidates who were less than ideal for a Fontan operation. Only one death occurred among the first 18 high-risk

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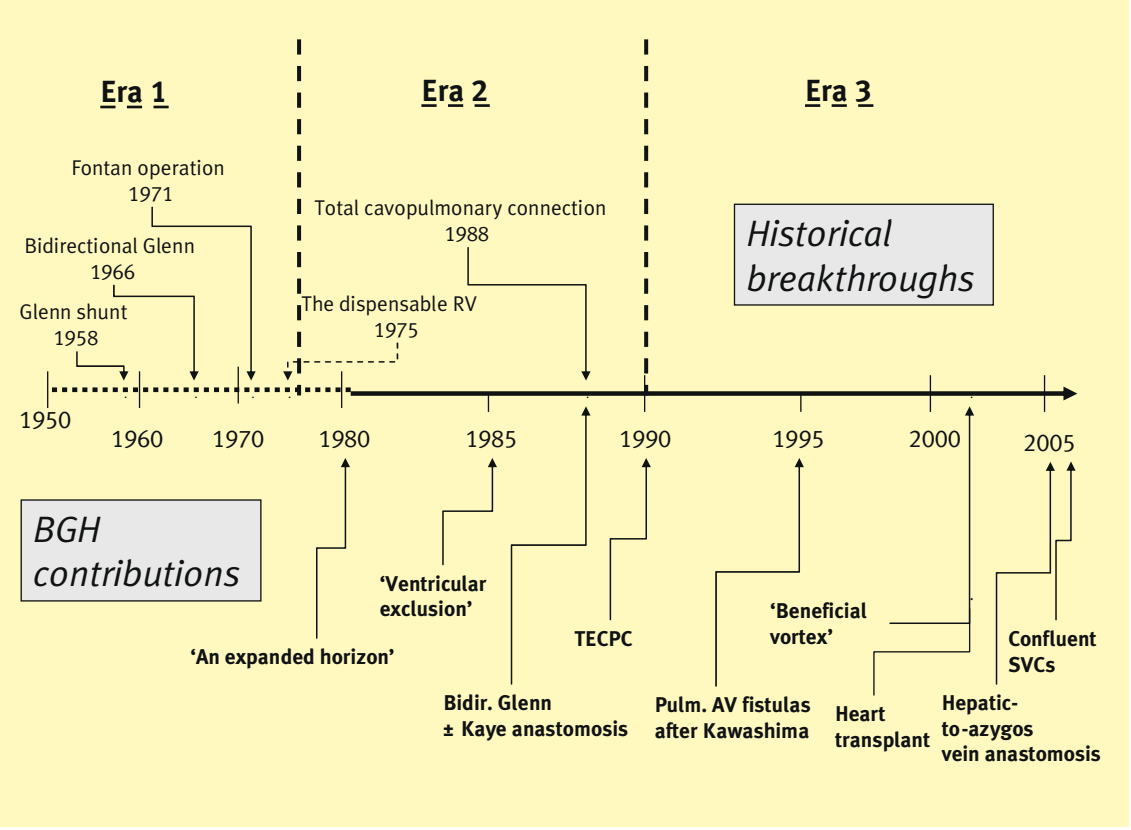

Figure 1 A schematic history of contributions to the surgical treatment of single-ventricle patients. AV, atrioventricular; BGH, Bambino Gesù Children's Hospital; RV, right ventricle; SVC, superior vena cava; TECPC, total extracardiac cavopulmonary connection.

patients, including four with interruption and azygos continuation of the inferior vena cava.[15] We postulated that the BCPA 'could avoid the sudden reduction of ventricular compliance that can be one of the reasons for failure of the Fontan principle in patients with a previous ventricular work load. The concept that the BCPA modulates the process of ventricular remodelling by partial volume unloading in single-ventricle patients has gained worldwide acceptance,[16] and this procedure is now universally considered to be an obligatory intermediate step preparatory to a Fontan operation.[17]

In the case of concomitant systemic outflow obstruction and consequent pressure overload, the degree of relative ventricular hypertrophy is even more prominent, further exacerbating the unfavourable geometric adaptation of the systemic ventricle to volume unloading at the time of Fontan repair. In 1986, we began to combine the BCPA with a *Damus–Kaye–Stansel anastomosis* to bypass the subaortic obstruction, obtaining simultaneous volume and pressure unloading.[18] The incorporation of the pulmonary artery root into the aortic reconstruction seemed preferable to either subaortic resection or the placement of a valved conduit between the ventricle and the ascending aorta due to the reduced risk of damaging the coronary arteries, the conducting tissue and the ventricular myocardium. A preliminary follow-up study on the first 23 patients showed no subaortic gradient at rest and a low end-diastolic ventricular pressure (a mean of 8 mmHg). Nine patients reached the Fontan stage with no hospital mortality, a major achievement considering the average worldwide early mortality of up to 40% for similar patients at that time.[19]

Concerning the potential use of BCPA as a definitive palliation, we were soon dissuaded because of the frequent occurrence of pulmonary arteriovenous malformations and systemic-to-pulmonary collaterals beginning about 2 years after the procedure.[20,21] This complication is possibly related to the lack of exposure of the pulmonary circulation to a putative hepatic or splanchnic vasoactive peptide carried only in the hepatic venous return, which is typically excluded in this cardiovascular arrangement.[22] Therefore, we no longer endorse the use of BCPA as a definitive surgical solution for high-risk candidates for a Fontan operation, nor do we encourage the addition of an alternative source of pulmonary blood flow, since this necessarily imposes a largely unpredictable volume load that potentially neutralises the process of ventricular remodelling sought by staging the Fontan operation.[23]

The extracardiac total cavopulmonary connection

In 1988, Mark de Leval introduced the revolutionary concept of a *total cavopulmonary connection*, i.e. a combination of a BCPA with a tubular *intra-atrial baffle* connecting the inferior vena cava to the pulmonary arteries (the *lateral tunnel technique*).[4] The rationale for this approach is that directing the systemic venous flows as concentric laminar wedges along cylindrical conduits assures a considerably more efficient kinetic energy preservation than do the stagnating, spinning swirls in the atrial reservoir of the old-fashioned atriopulmonary anastomosis. This hydraulic behaviour intuitively translates into increased functional performance and longevity of the total cavopulmonary

